

Coronary Cameral Fistula from Left Circumflex Artery to Left Atrium Identified by Three-Dimensional Transesophageal Echocardiography



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INTRODUCTION

A coronary cameral fistula is an anomalous connection between a coronary artery and a cardiac chamber. The etiology can be either congenital or iatrogenic. The clinical significance depends on the size and location of the fistula. Hemodynamic impairment can occur when the drainage site of the fistula is located in the left atrium or pulmonary vein, which in effect creates a left-to-left shunt. Continuous flow throughout the cardiac cycle can lead to volume overload, which may exacerbate heart failure and atrial and ventricular tachyarrhythmias.¹ This case presentation describes the rare echocardiographic visualization of an acquired coronary cameral fistula.

CASE PRESENTATION

A 74-year old Caucasian woman presented to our institution with dyspnea, fatigue, and edema. She had a history of rheumatic fever, severe mitral regurgitation, heart failure, atrial fibrillation, bioprosthetic valve replacement (27-mm Mosaic) in 2009, left atrial appendage ligation, and maze procedure. Physical examination findings were normal rhythm and normal heart sounds. She was admitted for symptoms of worsening heart failure with preserved ejection fraction. The patient underwent transthoracic echocardiography, which demonstrated a preserved ejection fraction of 55% to 60% and an increased mean transvalvular mitral gradient of 13 mm Hg compared with 8 mm Hg on transthoracic echocardiography 5 months prior. Transesophageal echocardiography (TEE) was recommended to evaluate the cause of elevated mitral valve gradients. Comprehensive TEE was performed using a Philips EPIC 7 (Philips Medical Systems, Andover, MA) with a transesophageal multiplane X8-2t omni sector array transducer. TEE demonstrated a normal ejection fraction, a mean mitral valve gradient of 7 mm Hg, and no overt evidence of thrombus or vegetation on the basis of three-dimensional echocardiography. In addition, there was abnormal flow noted on color Doppler flowing into the left atrium (Figure 1). The midesophageal (ME) short-axis view demonstrated a high-velocity color jet, which followed a tortuous course anteriorly.

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VIDEO HIGHLIGHTS

Video 1: Sweep of ME short-axis view demonstrating tortuous course of the circumflex artery.

Video 2: Three-dimensional en face view obtained from the ME view at 25° also aided in further characterization of the fistula in relation to other cardiac structures (AV and mitral valve).

View the video content online at www.cvcasejournal.com.

The continuous-wave Doppler sample gate was placed at the entry site and demonstrated continuous flow initiating at the onset of the QRS complex (Figure 2). A peak velocity was obtained from the continuous Doppler jet of 440 cm/sec. This was believed to be consistent with findings of a coronary cameral fistula. Coronary angiography was performed to further interrogate this suspicious jet, which confirmed the presence of a coronary cameral fistula between the left circumflex artery and the left atrium (Figure 3). In retrospect, the patient's previous transthoracic echocardiographic examinations did not reveal the presence of this fistula. Patient's shortness of breath improved with diuresis. Subsequently the patient was discharged with plans to return later for percutaneous treatment with a covered stent in the circumflex coronary artery. During follow-up office visits (over a 12-month period), the patient was found to be well compensated with minimal symptoms of heart failure, and she preferred not to undergo further intervention.

DISCUSSION

Coronary cameral fistulae are rare, with an incidence of 0.1% to 0.2%.² Blood bypasses normal capillary circulation and is shunted directly into cardiac chambers, great vessels, or other vascular structures.³ Congenital coronary cameral fistulae are more commonly reported as incidental findings, seldom require intervention, and are usually treated using medical therapy. Optimal management strategies are somewhat controversial, as there are mixed data regarding long-term risk for clinically "silent fistula."^{3,4} Potential complications of coronary cameral fistulae include mural thrombosis, rupture, aneurysm formation, and intimal rupture.^{1,5} Patients with coronary artery fistulae may present with symptoms including heart failure, coronary steal syndrome, endocarditis, pulmonary hypertension, and arrhythmias. Surgical or percutaneous interventions are indicated in patients who are symptomatic or present with coronary steal syndrome to alleviate worsening symptoms and improve clinical outcomes.⁶

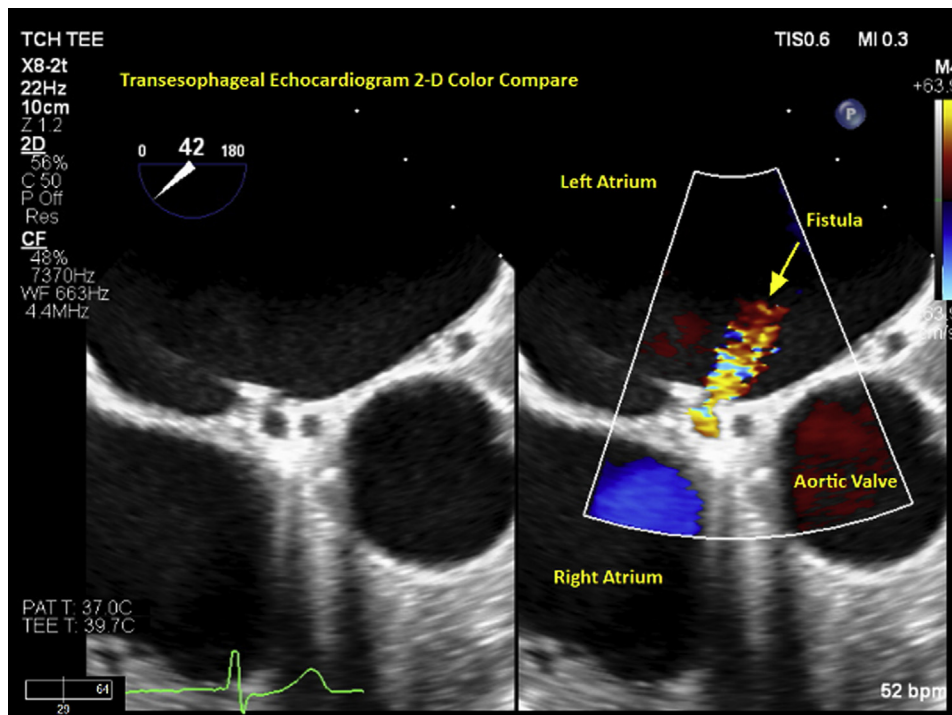


Figure 1 ME short-axis view demonstrating high-velocity color jet, which followed a tortuous course anteriorly.

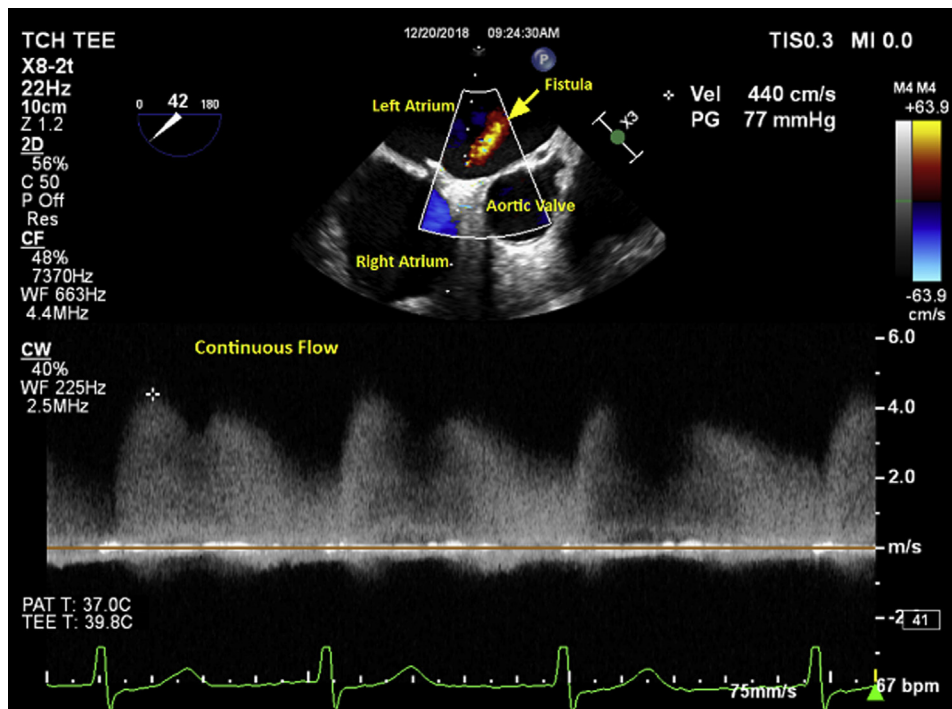


Figure 2 Continuous Doppler demonstrating continuous flow initiating at the onset of the QRS complex on the electrocardiogram.

Percutaneous therapeutic options include vascular plugs, occlusion coils, and covered stents.

Coronary cameral fistulae most frequently originate from the right coronary artery. However, in our case we describe a less common origin from the left circumflex coronary artery, which was likely acquired following mitral valve surgery. Given the close proximity of the left circumflex coronary artery to the atrioventricular groove,

direct injury or distortion of the circumflex has been reported in up to 2% of cases following mitral valve surgery.⁷ Of note, the proximal third of the circumflex has the shortest distance between the mitral annulus, making it more susceptible to iatrogenic injury during mitral valve surgery.^{8,9}

Although infrequent and subtle in its presentation, coronary cameral fistulae can be detected by TEE, as previously reported.¹⁰

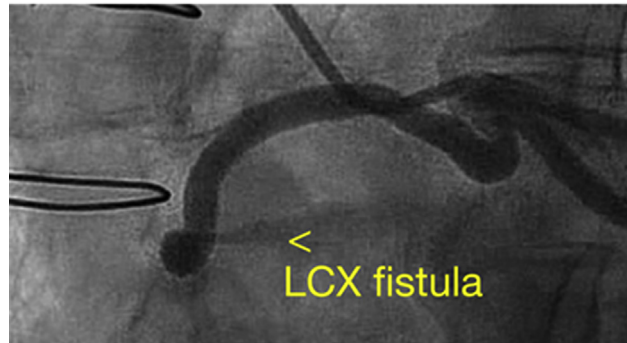


Figure 3 Coronary angiogram demonstrating coronary cameral fistula between the left circumflex coronary artery (LCX) and left atrium, consistent with findings on TEE.

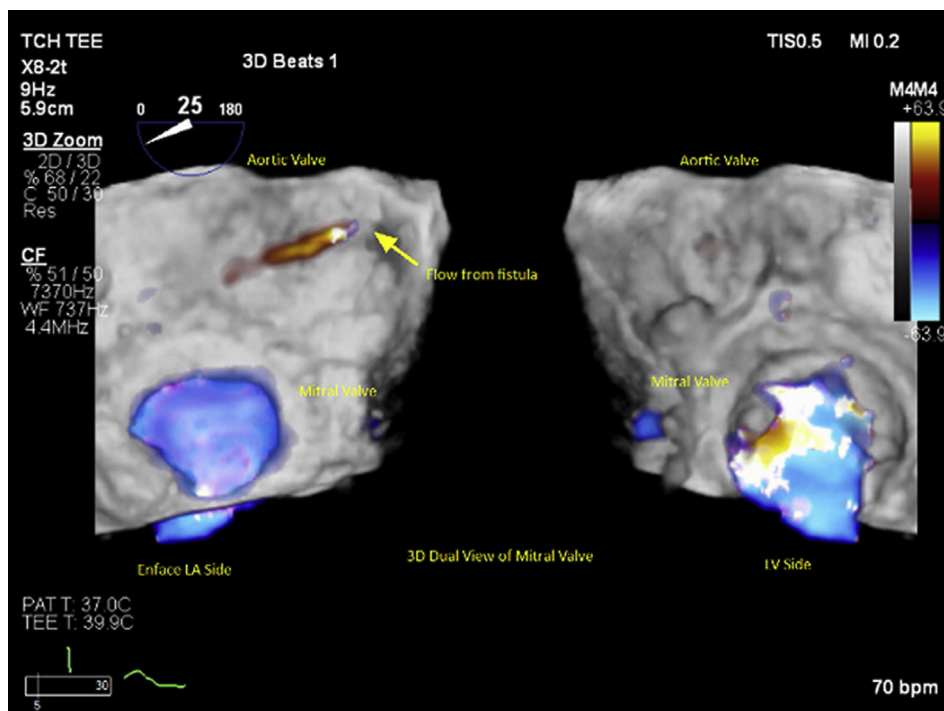


Figure 4 Three-dimensional en face view obtained from the ME view at 25° also aided in further characterization of the fistula in relation to other cardiac structures (AV and mitral valve). Dual view demonstrating simultaneous color flow emanating from the fistula (viewed from both the atrial and ventricular aspects at the level of the mitral valve).

Two-dimensional TEE and color Doppler interrogation demonstrated a suspicious jet flowing into the left atrium. Three-dimensional echocardiography further clarified the anatomic characteristics of the fistula. Although angiography is considered the gold standard for the diagnosis of coronary cameral fistula, our case study demonstrates that this anomaly can be clearly detected using noninvasive techniques. The left circumflex coronary artery can be imaged in 90% of patients from the ME aortic valve (AV) short-axis view and is best at demonstrating the proximal portion of the artery. The proximal and distal portions of the left circumflex can also be imaged from the ME AV long-axis view.¹¹ In our case the proximal portion of the left circumflex coronary artery was visualized from the ME AV short-axis view at 45°, which followed a tortuous course (anteriorly) emptying into the left atrium (Figure 1, Video 1). A sweep revealed the origin of the high-velocity color jet to be at the takeoff of the

left circumflex coronary artery (Video 1). A three-dimensional en face view obtained from the ME view at 25° also aided in further characterization of the fistula in relation to other cardiac structures (AV and mitral valve; Figure 4, Video 2). TEE can help locate the origin of fistulae and provide visualization of posterior cardiac structures, particularly in patients with complex anatomy.¹²

CONCLUSION

This case demonstrates a coronary cameral fistula, which although infrequent can be the result of iatrogenic injury following cardiac surgery. This case also highlights the utility of noninvasive echocardiography, which can be used in the initial detection of this defect. Additionally, the optimal TEE strategy should include a

comprehensive protocol comprising two-dimensional, three-dimensional, color, and spectral Doppler imaging.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2020.05.013>.

REFERENCES

1. Buccheri D, Chirco PR, Geraci S, Caramanno G, Cortese B. Coronary artery fistulae: anatomy, diagnosis and management strategies. *Heart Lung Circ* 2018;27:940-51.
2. Cebi N, Schulze-Waltrup N, Fromke J, Scheffold T, Heuer H. Congenital coronary artery fistulas in adults: concomitant pathologies and treatment. *Int J Cardiovasc Imaging* 2008;24:349-55.
3. Gowda RM, Vasavada BC, Khan IA. Coronary artery fistulas: clinical and therapeutic considerations. *Int J Cardiol* 2006;107:7-10.
4. Latson LA. Coronary artery fistulas: how to manage them. *Catheter Cardiovasc Interv* 2007;70:111-8.
5. Challoumas D, Pericleous A, Dimitrakaki IA, Danelatos C, Dimitrakakis G. Coronary arteriovenous fistulae: a review. *Int J Angiol* 2014;23:1-10.
6. Dodge-Khatami A, Mavroudis C, Backer CL. Congenital heart surgery nomenclature and database project: anomalies of the coronary arteries. *Ann Thorac Surg* 2000;69(4 suppl):S270-97.
7. Aybek T, Risteski P, Miskovic A, Simon A, Dogan S, Abdel-Rahman U, et al. Seven years' experience with suture annuloplasty for mitral valve repair. *J Thorac Cardiovasc Surg* 2006;131:99-106.
8. Grande AM, Fiore A, Massetti M, Viganò M. Iatrogenic circumflex coronary lesion in mitral valve surgery: case report and review of the literature. *Texas Heart Inst J* 2008;35:179-83.
9. Pessa C, Gomes W, Catani R, Prates J, Buffolo E. Anatomical relationship between the posterior mitral valve annulus and the coronary arteries. Implications to operative treatment. *Braz J Cardiovasc Surg* 2004;19:372-7.
10. Iida R, Yamamoto T, Suzuki T, Saeki S, Ogawa S. The usefulness of intraoperative transesophageal echocardiography to identify the site of drainage of coronary artery fistula. *Anesth Analg* 2005;101:330-1.
11. Ender J, Singh R, Nakahira J, Subramanian S, Thiele H, Mukherjee C. Echo didactic: visualization of the circumflex artery in the perioperative setting with transesophageal echocardiography. *Anesth Analg* 2012;115:22-6.
12. Almeida J, Lobo A, Casanova J, Torres JP, Gomes MR. Transesophageal echocardiography on the presurgical and postsurgical evaluation of a coronary arteriovenous fistula. *J Am Soc Echocardiogr* 1997;10:224-7.